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MRI of Intimal Sarcoma of the Pulmonary Arteries

Pia K. Schuler, MD; Achim Weber, MD; Peter K. Bode, MD;
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A 38-year-old woman presented with left lower chest pain. At the age of 3 years, a commissurotomy of the pulmonary valve and infundibulectomy was performed to correct severe infundibular pulmonary stenosis. The initial thoracic CT diagnosed bilateral pulmonary embolism. Echocardiography revealed a filiforme mass in the pulmonary artery 33 mm in length, floating in the right outflow tract, suggesting a pulmonary sarcoma. In addition, a moderate pulmonary valve insufficiency with mild stenosis was diagnosed. Cardiac magnetic resonance (CMR) confirmed the mass in the left pulmonary artery (PA) with floating parts and a small mass in the right PA (Figure 1) and provided complementary findings identifying the mass as tumor, differentiating it from chronic thromboembolism. In particular, perfusion CMR¹ demonstrated blood flow in the mass (Figure 2A and 2B), and late contrast-enhanced CMR revealed high- and low-signal territories in the tumor suggesting heterogenous cell-rich and cell-depleted components of the mass (Figure 3).²

Histology as shown in Figure 4 showed the mass to be an intimal sarcoma with widespread vascularization with cell-rich and myxoid, cell-depleted regions.

The patient underwent an open surgical resection of the pulmonary mass including repair of the PA and pulmonary valve replacement. An additional chemotherapy with adriamycin/Holoxan and uromitexan was started 4 months later, and a lobectomy of the right upper pulmonary lobe was performed for metastatic disease. One year after being diagnosed, the patient is still alive and has started working again.

The clinical presentation and radiological features may mimic pulmonary embolism.³ The CMR criteria suggesting tumor rather than emboli are (1) detectable tissue perfusion and (2) heterogenous enhancement on late CMR imaging resulting from differences in tumor cellularity.²

Intimal sarcoma is a rare, highly malignant neoplasm that typically arises from the central pulmonary arteries. The prognosis must be regarded as extremely poor. Most patients die within months of right heart failure secondary to outflow obstruction or distal (thrombo) embolism. The main treatment for sarcoma is aggressive surgical resection, but, even after heart and lung transplantation, recurrent metastases in noncardiac organs occurred in half of

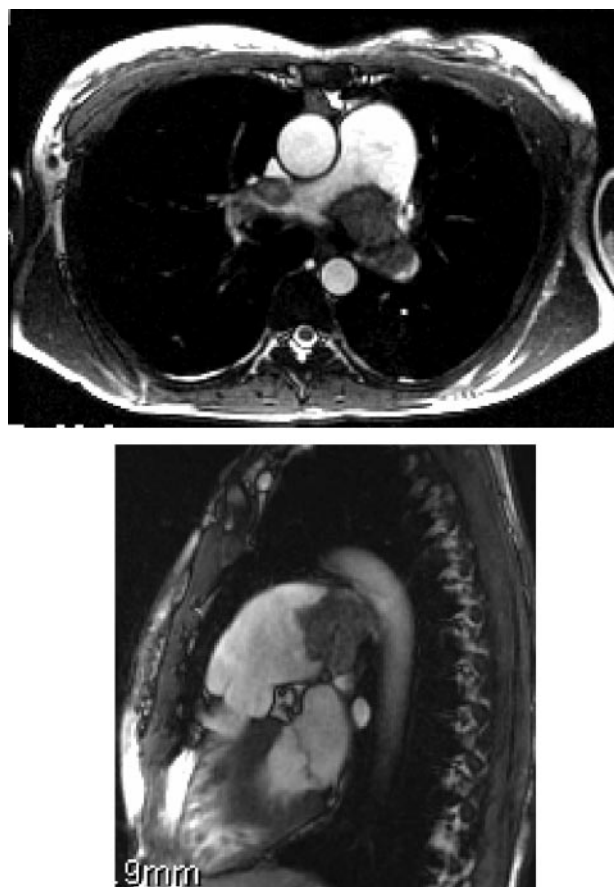


Figure 1. Mass in the left and right PA in axial (A) and sagittal (B) views (steady-state free precession) are shown.

the patients. The role of adjuvant radiation and chemotherapy remains unclear.^{3,4}

CMR provides important information, readily distinguishing tumors such as intimal sarcoma from thrombotic masses in the pulmonary arteries, which are notoriously difficult to discriminate with conventional diagnostic approaches.

Disclosures

None.

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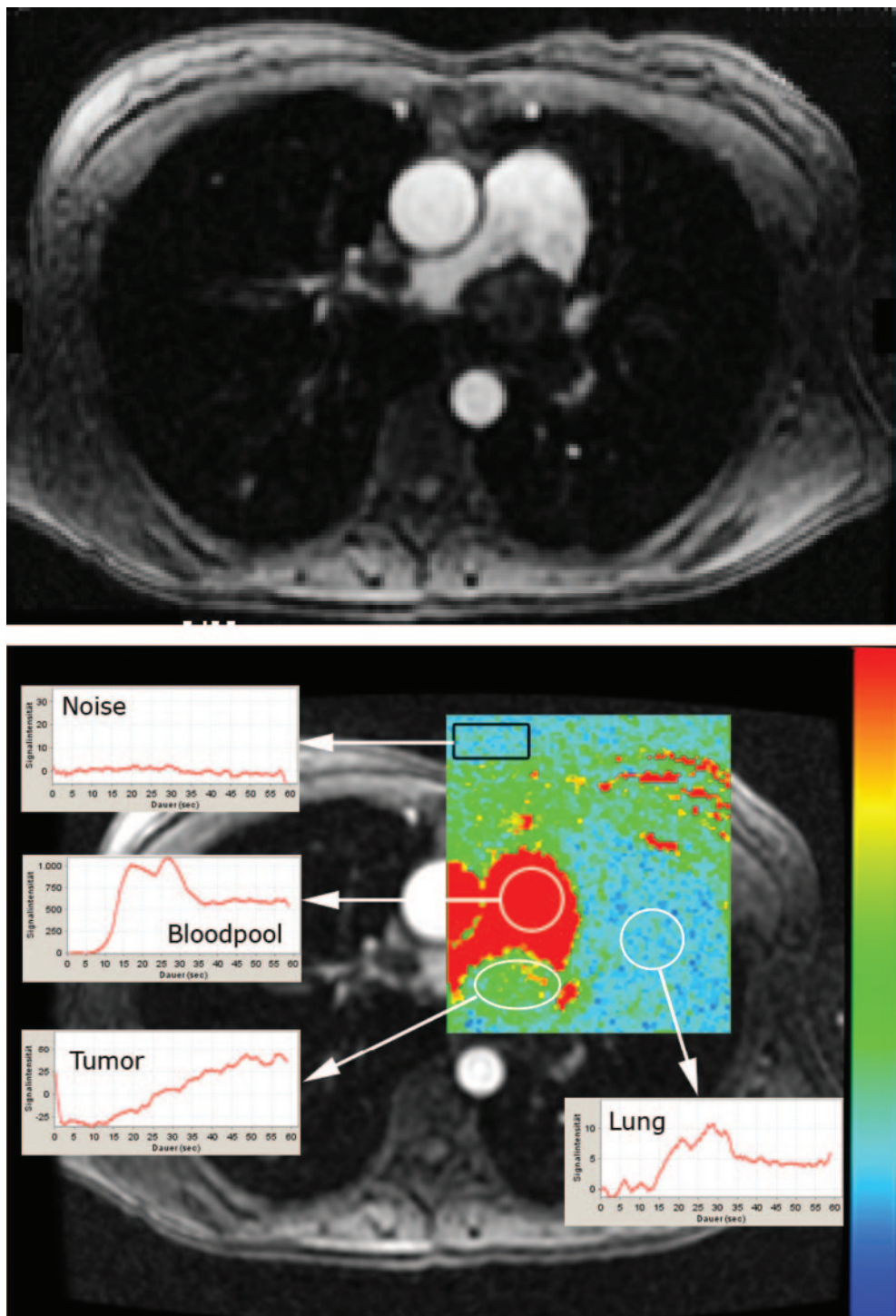


Figure 2. Perfusion CMR shows signal increase during first pass of contrast medium through the mass at peak bolus (A) and represented as upslope map and signal-intensity-time curves (B).



Figure 3. Late gadolinium-enhanced CMR identifies irregular enhancement patterns in the tumor (the optimal TI was set to null normal myocardium).

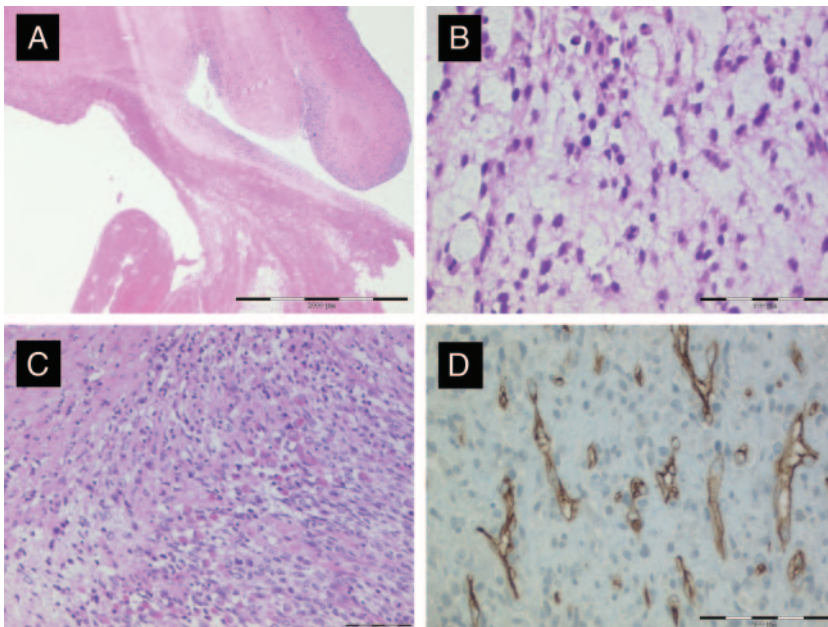


Figure 4. A, Vessel wall (upper part) with polypoid tumor formations and adjacent tumor necrosis and thrombotic material (lower part) (hematoxylin and eosin staining; scale bar, 2000 μm). B, Myxoid area with low cellular density and mitotic activity (hematoxylin and eosin staining; scale bar, 100 μm). C, Rhabdoid features (hematoxylin and eosin staining; scale bar, 200 μm). D, CD34 immunostaining highlights numerous small vascular clefts (scale bar, 100 μm).

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